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CASE REPORT OPEN ACCESS

Diaphragmatic Solitary Fibrous Pleura Tumor Masquerading as Lung Tumor- A Case Report

Anumenechi N[1ID], Jibrin P[2], Epepe U[1ID]

[1]Cardiothoracic Unit, Department of Surgery, National Hospital Abuja, Nigeria
[2]Department of Histopathology, National Hospital Abuja, Nigeria

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Abstract
Background: Solitary pleural fibrous tumours of the diaphragm are rare and can be a diagnostic dilemma.
Case presentation: A 49-year-old female presented to our clinic with a history of chronic cough with associated difficulty in breathing. Chest findings were dull percussion notes as well as reduced breath sounds on the left lower lung zone. Chest CT scan showed a left-sided intra-thoracic density with compression of the left lower lung lobe. She had a thoracotomy with the excision of a left diaphragmatic pleura tumour.
Conclusion: This case intends to highlight that, though primary diaphragmatic pleura tumours are rare, they may present with non-specific symptoms and should be considered in evaluating patients with intra-thoracic tumours.

Keywords: Diaphragmatic tumour, Diaphragmatic fibroma, Pleural fibroma, Parietal pleural tumour

Background

Solitary diaphragmatic pleura tumors are tumors arising from the parietal pleura overlying the diaphragm, they may be benign or malignant. Solitary diaphragmatic pleura tumours are rare (1).

Pleura-based masses can be primary or secondary in origin, most are metastases from the lungs, thymus, or extra-thoracic malignancies like breast or prostate cancer (2). However, primary pleura tumours do occur and can also be benign or malignant. The benign tumours are mostly fibrous (3).

The primary pleura-based tumours originate from the parietal pleura layer of the thoracic cavity, and they pose significant pathologies by compressing adjacent structures with wide-ranging loco-regional clinical features. They can also present symptoms attributable to para-neoplastic phenomena like persistent hypoglycemia in Doege-Potter syndrome (4, 5).

This disease is rare, with occasional case reports occurring across all age groups, from childhood to old age (6). Almost all benign pleura tumours are solitary, they are commonest over the inner layer of the rib cage. Their occurrence over the diaphragm is less common and, in that location, can be confused with diaphragmatic tumors which in themselves can also be primary or secondary.

Clinical features depend on organ infiltration and can be chest pain, breathlessness, cough, hiccups, and fainting spells among other symptoms.

Diagnosis needs a high index of suspicion and appropriate diagnostic investigations. Biopsies for histologic evaluation and immuno-histochemistry are very important.

We report herein the case of a patient with a histologically diagnosed solitary fibrous tumour of the diaphragmatic pleura who had complete resection of the tumour.

Case presentation

The patient was a 49-year-old female, who presented to our clinic with a productive cough of eleven months duration with associated difficulty in breathing, worse on exertion. The cough was productive of whitish sputum, which was occasionally cream-coloured.

Correspondence:
Anumenechi, Ndubuisi
Cardiothoracic Unit, Department of Surgery,
National Hospital
Abuja, Nigeria
+2348067795916, cardio_dr@yahoo.com

On physical examination, she was not cyanosed and had no digital clubbing, no peripheral lymphadenopathy, or pedal oedema. There were dull percussion notes as well as reduced breath sounds on the left lower lung zone.
Chest X-ray showed a homogenous opacity on the left lower lung zone. A chest CT scan revealed a homogenous mass measuring 16.8 x 12.6 cm in the left chest compressing the lower lung lobe **(Figure 1)**. CT-guided lung biopsy showed idiopathic pulmonary fibrosis, lung function test revealed a restrictive disorder; FVC of 76% and FEV1/FVC of 82%.

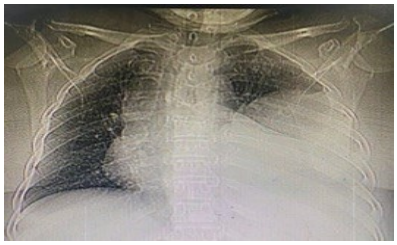


Figure 1a: Preoperative chest CT scan (a) coronal view: well-circumscribed density occupying two-thirds of the left hemithorax, with contralateral mediastinal shift

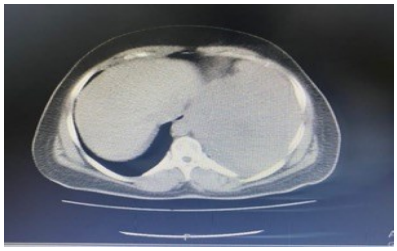
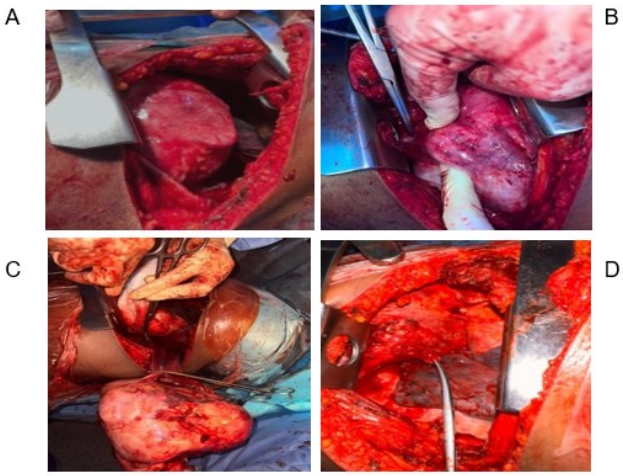


Figure 1b: Axial view of mass seen in Figure 1a

The provisional diagnosis was a left intra-thoracic tumour with consideration of a left lower lung lobe tumour. She had left postero-lateral thoracotomy with intra-operative findings of a huge fibrous tumour measuring 20 x 20 cm and weighing 5 kg. The tumor was attached to the dome of the left hemi-diaphragm with a base of 10cm, and compressing the left lung lower lobe There were dense adhesions between the tumor and the left lower lobe. The tumour was resected, with a sleeve of the parietal pleura overlying the fibrous part of the left diaphragm **(Figure 2 a-d)**. A chest tube was placed for drainage with an estimated blood loss of 100mls.



a: tumour shown inferiorly in the wound, sitting on the left diaphragm, exposed via a left posterolateral thoracotomy
b: tumor dissection
c tumour delivered out of the chest, artery forceps clamping dense adhesion
d Tumor bed after excision-tip of artery forceps on left hemidiaphragm, collapsed left lung is seen deep to the forceps

Figure 2: Intraoperative images

Post-operatively she was nursed in the intensive care unit for one day, she improved and was discharged on the fifth postoperative day.

Histology of the resected tumour revealed a mesenchymal tumour composed of haphazardly arranged spindle to oval-shaped cells with focal

areas of hyalinization and myxoid changes. Few papillary-sized blood vessels are also seen with vague areas of cartwheel appearance. No recognizable pulmonary parenchyma tissue epithelial tissue or cellular atypia was seen. The conclusion was a solitary fibrous tumour (Figure 3 a-d).

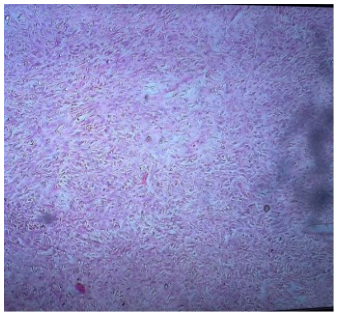


Figure 3a: H&E stain X10 magnification: mesenchymal tumour composed of haphazardly arranged spindle oval cells with focal areas of hyalinization and myxoid changes. Few papillary-sized blood vessels are also noted. Vague areas of cartwheel appearance. No recognizable pulmonary parenchymal tissue or epithelia seen, no atypia. Conclusion – solitary fibrous tumour

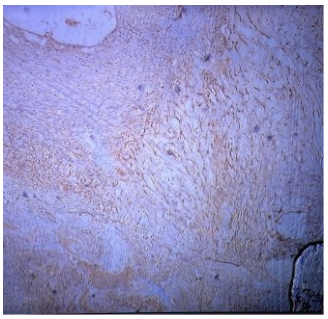


Figure 3b: EMA stain: positive magnification X10: moderate patchy membranous to cytoplasmic positivity

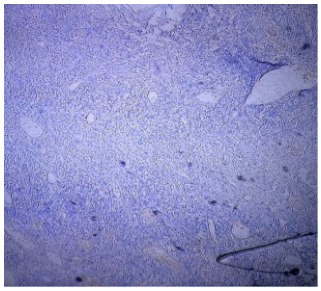


Figure 3c: Desmin stain: negative magnification X10

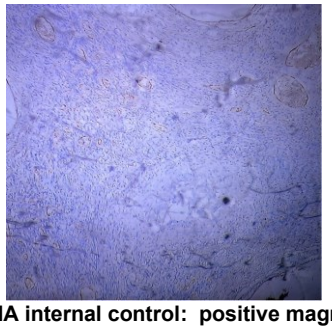


Figure 3d: SMA internal control: positive magnification X10

A 2-week and 1-month follow-up visit showed resolution of symptoms and satisfactory left lower lung lobe re-expansion (Figure 4).

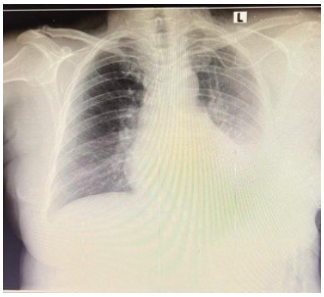


Figure 4a: 2 weeks post-op Chest X-ray; tumor resected

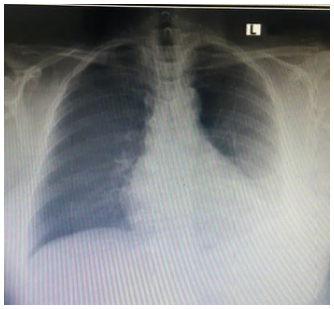


Figure 4b: 1-month post-op Chest X-ray; mild left pleural effusion

Discussion

The incidence of benign and malignant primary tumours of the diaphragm pleura is similar (7). The most common benign pleural tumours are cysts and lipomas (8), while the most common primary malignant tumours of the diaphragm include rhabdomyosarcoma and fibrosarcoma. Solitary fibrous pleural tumors are rare, Zarrouk *et al* described 5 cases over 11 years (3). Secondary tumours of the diaphragm with origin in the thoracic or abdominal cavity occur more frequently than the primary tumours. Yanagiya *et al* reported five cases described in the literature (9). Distinguishing between a lung and a diaphragmatic tumour can be difficult.

Symptoms can be varied and bizarre, with a reported case misdiagnosed as a psychiatric disease and tuberculosis (10). The presentation of our reported case was mainly pulmonary symptoms and pain due to atelectasis following lung compression by the tumour. However, she did not have recurrent or persistent hypoglycemia of Doege-Potter syndrome, which is due to the secretion of insulin-like growth factor by mesothelial fibrous tumours (5). The adhesions found intra-operatively could be attributed to the recurrent pneumonia from atelectasis.

The histology showed a benign fibrous tumour. Identifying the origin as either diaphragmatic muscle or the overlying parietal pleura required immunohistochemistry, the positive EMA showed pleura origin, while the negative Desmin and SMA ruled out diaphragmatic muscle origin (11). (Figure 4 a-b).

The presentation of the patient with a diaphragmatic tumour is variable depending on age at presentation, size of mass, involvement of adjacent organs, metastatic disease, and tumour histology. Approximately 50% of cases are asymptomatic, discovered either incidentally on radiographs or at surgical exploration performed for unrelated reasons (9). Large masses may give rise to compression of adjacent structures such as the lung causing cough, dyspnea, hemoptysis, or lower extremity oedema if the venous return is compromised.

Minimally invasive surgical options have been reported in the surgical treatment of pleural or diaphragmatic fibromas, like VATS and laparoscopic surgery is useful, especially in small-size tumors. Kamiya *et al* reported a laparoscopic resection of diaphragmatic schwannoma (13).

Recurrence is high in solitary fibrous tumours, even in benign cases (13) and lifelong follow-up is recommended (14).

Conclusion

Solitary fibrous diaphragmatic pleural tumours are rare, with challenging preoperative diagnosis and very diverse and non-specific clinical features. A high index of suspicion of this tumour should be considered in patients presenting with features suggestive of lung lesions.

List of Abbreviations

CT: Computed Tomography
FEV1: Forced Expiratory Volume in 1 second
FVC: Forced Vital Capacity
EMA: Epithelial Membrane Antigen
SMA: Smooth Muscle Actin

Declarations

Ethical approval and consent to participate

The patient and relatives voluntarily agreed for the case to be published, her anonymity was preserved and no harm came to her because of the case report

Consent for publication
All authors gave consent for publication of the work under the Creative Commons Attribution-Non-Commercial 4.0 license.

Availability of data and materials
All essential data supporting the findings of this case are available within the article. Additional data are available upon request from the corresponding author.

Competing interests
The authors declare no conflict of interest.

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Authors' contributions
All the authors fully participated in the conception of the idea, literature review and approved the final draft of the article

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